Radiology Case Reports 20 (2025) 2184–2188

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Partial anomalous pulmonary venous connection and an incidental aberrant right subclavian artery: A rare case report ☆

Ashish Khadka, MD[a], *Aakash Neupane, MBBS*[b,*], *Pramodman Singh Yadav, MBBS*[b], *Leeza Shah, MD*[c], *Abinash Dev, MBBS*[b], *Sabin Ghale, MBBS, MBBS*[b], *Amisha Karki, MBBS*[d]

[a] *Department of Radiodiagnosis and Imaging, National Academy of Medical Sciences, Kathmandu, Nepal*
[b] *Department of Internal Medicine, B.P. Koirala Institute of Health Sciences, Dharan, Nepal*
[c] *Department of Internal Medicine, Chitwan Medical College, Nepal*
[d] *Department of Internal Medicine, Bharatpur Hospital, Bharatpur, Nepal*

ARTICLE INFO

Article history:
Received 13 September 2024
Revised 16 December 2024
Accepted 6 January 2025

Keywords:
Congenital heart diseases
Pulmonary vein anomalies
Cardiology
Dyspnea
Computed tomography

ABSTRACT

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital heart defect where some pulmonary veins drain into the right atrium instead of the left. We report a case of a 20-year-old female with worsening dyspnea and intermittent chest pain. Imaging revealed cardiomegaly and pulmonary hypertension on chest X-ray. Contrast-enhanced CT showed normal right pulmonary vein drainage but anomalous drainage of the left pulmonary veins into the left brachiocephalic vein. An incidental finding of an aberrant right subclavian artery (ARSA) was also noted. The patient underwent successful surgical correction of PAPVC, with no postoperative complications, though long-term follow-up was unavailable. PAPVC is often asymptomatic but can lead to right-sided heart failure and pulmonary hypertension if untreated. ARSA, a rare anomaly, was not clinically significant in this case. This report emphasizes the role of advanced imaging in diagnosing rare anomalies like PAPVC and ARSA, where early detection and intervention are crucial for preventing complications like right ventricular dysfunction and arrhythmias.

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital heart defect where some pulmonary veins return blood to the right atrium instead of the left. Typically, only 1 pulmonary vein is affected, though all veins from 1 lung can be anomalous in rare cases [1]. This condition causes some pulmonary venous blood to enter the systemic venous circulation. PAPVC is found in 0.4%-0.7% of autopsy cases, though

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: aakashneupane16@gmail.com (A. Neupane).

https://doi.org/10.1016/j.radcr.2025.01.027

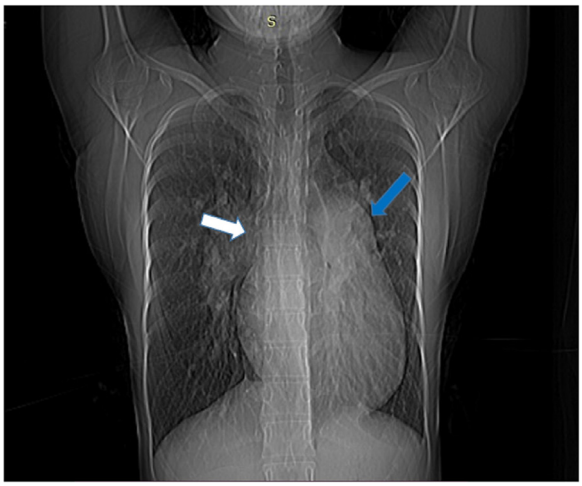

Fig. 1 – Chest X-ray showing increased cardiothoracic ratio with prominent Pulmonary Bay with enlarged right descending pulmonary artery.

many of these may be asymptomatic, making the true incidence lower [2]. It is associated with atrial septal defects (ASD) in about 10% of cases. PAPVC creates a left-to-right shunt, similar to ASD or ventricular septal defects (VSD), and is more commonly found on the right side, often detected incidentally via imaging. At least 50% of anomalous venous flow is usually required for clinical significance [3]. Understanding normal pulmonary venous drainage patterns is crucial for diagnosis. We present a rare case of PAPVC where the left pulmonary vein drains into the brachiocephalic vein with an incidental finding of aberrant origin of the right subclavian artery.

Case presentation

A 20-year-old female presented with a history of progressively increasing dyspnea over 1 week accompanied by intermittent chest pain. There was no history of fever, cough, malaise, or cyanosis. The patient had a history of repeated hospital admission for shortness of breath. Lab investigations were within normal limits.

Imaging studies

Frontal Chest Xray PA view: Frontal Chest Radiograph (PA View) revealed an increased cardiothoracic ratio suggesting cardiomegaly. The pulmonary bay was prominent with convexity outwards and a lateral border of convexity lying outside a tangent drawn between the Aortic knuckle and left heart border (suggesting an enlarged pulmonary trunk). The right hilum was bulky with an enlarged right descending pulmonary artery. All these features are suggestive of pulmonary artery hypertension as a result of increased blood flow into the right side of the heart (Fig. 1).

CECT chest findings

A contrast-enhanced Computed Tomography imaging of the chest revealed normal venous drainage of the right superior and inferior pulmonary veins into the left atrium (Figs. 2A-C). However, left superior and inferior pulmonary veins do not drain into the left atrium and instead continue cranially (Figs. 3A and B). The superior and inferior pulmonary veins unite to form a common trunk and continue cranially (Figs. 4A and B). The common trunk of the left superior and inferior pulmonary vein finally drains anomalously into the left brachiocephalic vein (Fig. 5). The findings can be further corroborated in this coronal view (Fig. 6).

CT scan also revealed an incidental finding of an aberrant subclavian artery. Axial section CT scan showed 4 vessels originating from the aortic arch instead of the normal 3 vessels with an absence of brachiocephalic trunk with separate origin of the right common carotid artery and right subclavian artery from the aortic arch (Figs. 7A and B). There was an anomalous course of the aberrant right subclavian artery posterior to the esophagus (Figs. 8A and B).

Management

The patient was prepped for surgery with an uneventful preoperative period. The patient underwent open PAPVC repair and was monitored in the Cardiac Care Unit (CCU) postoperatively and was discharged after 1 week. Long-term follow-up data is not available as the patient was lost to follow-up.

Discussion

The failure of normal pulmonary vein embryological development can result in a range of congenital anomalies, from normal variations to partial anomalous pulmonary venous connection (PAPVC) and total anomalous pulmonary venous connection (TAPVC) [1]. These conditions may present in isolation, often accompanied by an atrial septal defect (ASD), or as part of complex congenital heart diseases (CHD). PAPVC is a congenital cardiac anomaly with no established cause, potentially having a multifactorial origin that includes a genetic component. It has been observed in individuals with Turner syndrome, though no definitive genetic predisposition or common teratogens (such as drugs or infections) have been implicated in its development.

Left-sided PAPVC is significantly less common than right-sided PAPVC, with the former affecting the upper lobe drainage more frequently than the entire lung drainage. Typically, the anomalous pulmonary vein drains via a vertical vein into the innominate vein, although rarer drainage patterns may involve the left subclavian vein, persistent left superior vena cava, coronary sinus, or directly into the right atrium. The venous connection patterns in PAPVC vary, and mixed connections—where veins from each lung drain into different compartments—are also reported. Hemodynamically, right atrial pressure is generally lower than left atrial pressure, making the anomalously draining side more prone to venous return [4]. Most PAPVC cases are discovered incidentally since patients are often asymptomatic or present with mild symptoms. Cyanosis, when present, tends to manifest later in life due to increased pulmonary vascular resistance and pulmonary hypertension [5].

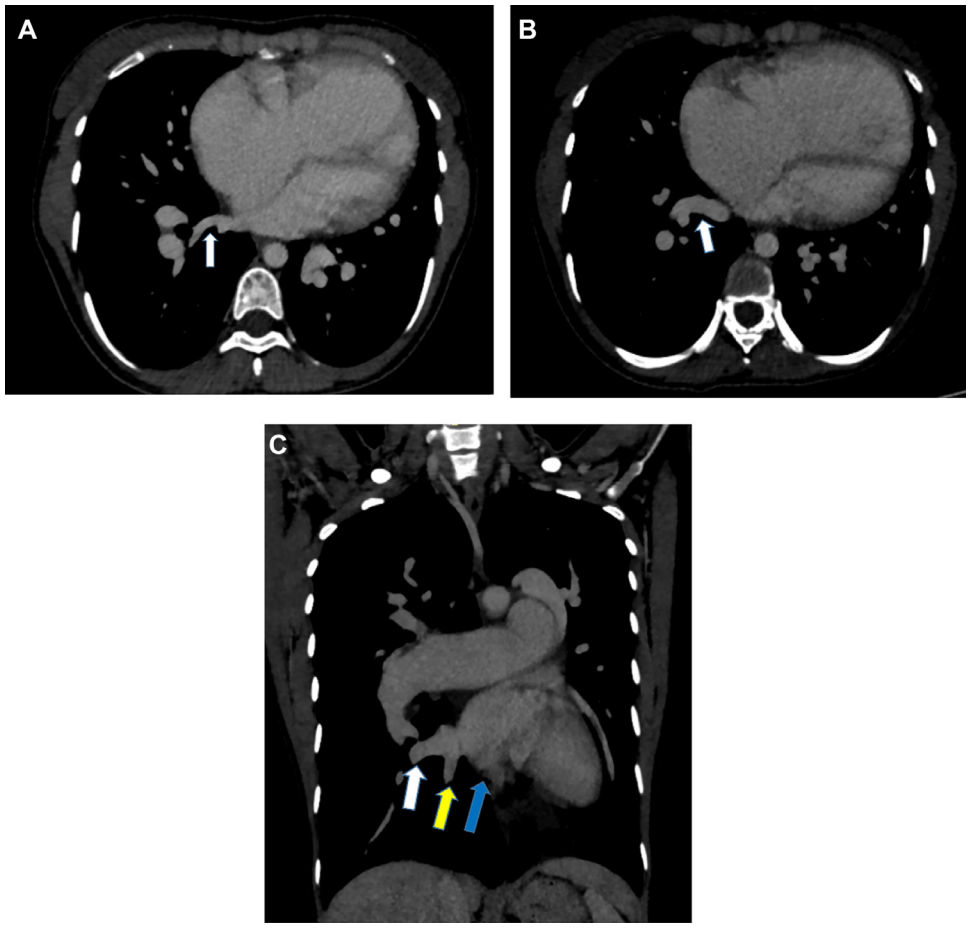

Fig. 2 – (A) Normal venous drainage of the right superior pulmonary vein (White arrow) into the left atrium (B) Normal venous drainage of the right inferior pulmonary vein (White arrow) into the left atrium (C) Coronal CT showing normal drainage of right pulmonary veins into right atrium with White arrow showing Right superior pulmonary vein, yellow arrow showing right inferior pulmonary vein and blue arrow showing left atrium.

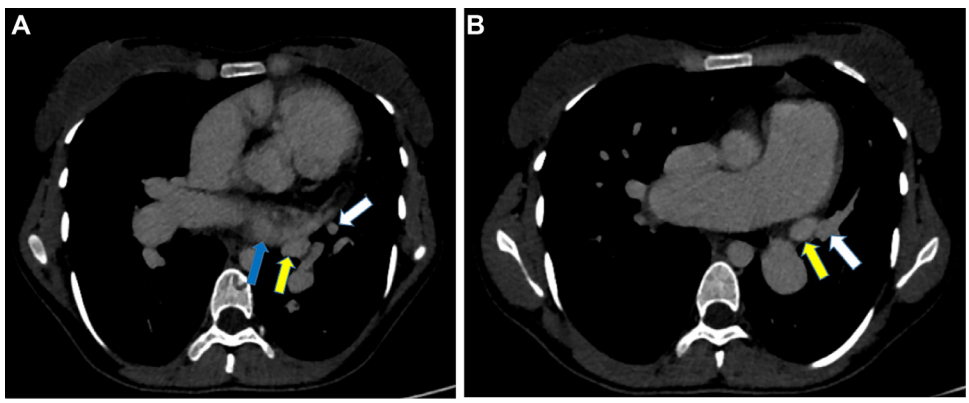

Fig. 3 – (A and B) Cranial continuation of right superior pulmonary vein (White Arrow) and inferior pulmonary vein (Yellow Arrow) without draining into left atrium (Blue Arrow).

Isolated PAPVC is frequently asymptomatic and may remain undiagnosed until adulthood, as seen in this report. The clinical presentation and progression of PAPVC largely depend on the shunt fraction, which is influenced by the number of pulmonary veins draining into the right heart. A single anomalous vein is typically not hemodynamically significant unless accompanied by other left-to-right shunts, such as ASDs. If left untreated, prolonged PAPVC can lead to right-sided heart volume overload, tricuspid regurgitation, arrhythmias, pulmonary hypertension, and right ventricular dysfunction [6]. Diagnostic approaches for PAPVC often involve echocardiography, though it may be limited by its

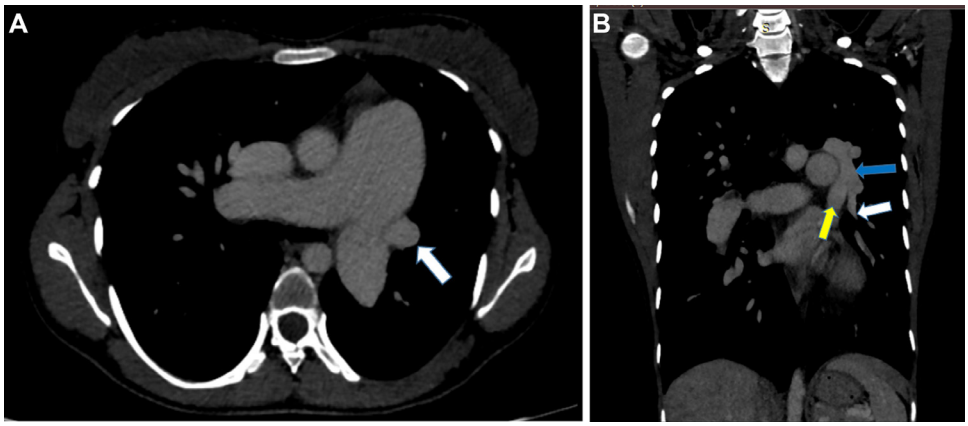

Fig. 4 – (A) Superior and inferior left pulmonary veins unite to form a common trunk (White arrow) – sagittal view (B) Coronal CT showing left superior pulmonary vein (White Arrow) and left inferior pulmonary vein (White Arrow) uniting to form a common trunk (Blue Arrow).

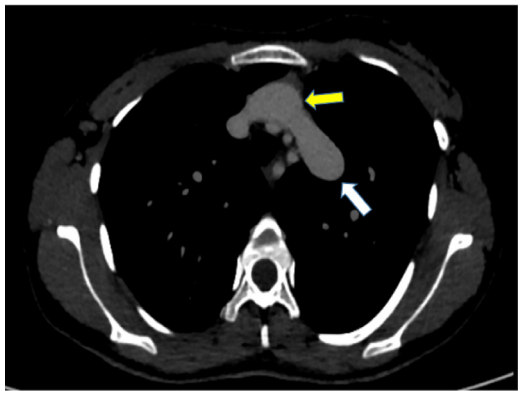

Fig. 5 – Common pulmonary vein trunk (White Arrow) draining anomalously into left brachiocephalic vein (Yellow Arrow).

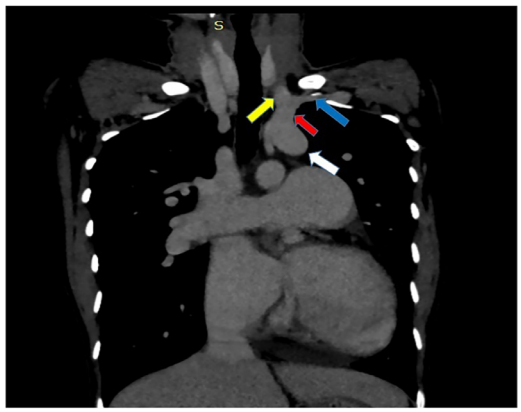

Fig. 6 – Coronal view of anomalous drainage of common trunk of left superior and inferior pulmonary veins (White Arrow) into brachiocephalic vein (Red Arrow). Other venous structures draining into left brachiocephalic vein- yellow arrow: Left internal jugular vein, blue arrow: Left subclavian vein.

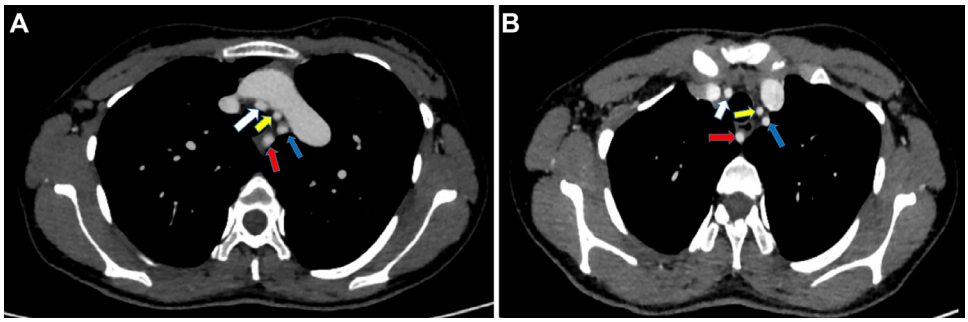

Fig. 7 – (A and B) Image showing absence of right brachiocephalic trunk and separate origin of right common carotid and right subclavian artery from aortic arch, red arrow: Aberrant course of right subclavian artery posterior to the esophagus (ARSA), White arrow: Right common carotid artery. Yellow arrow: Left common carotid artery, blue arrow: Left subclavian artery.

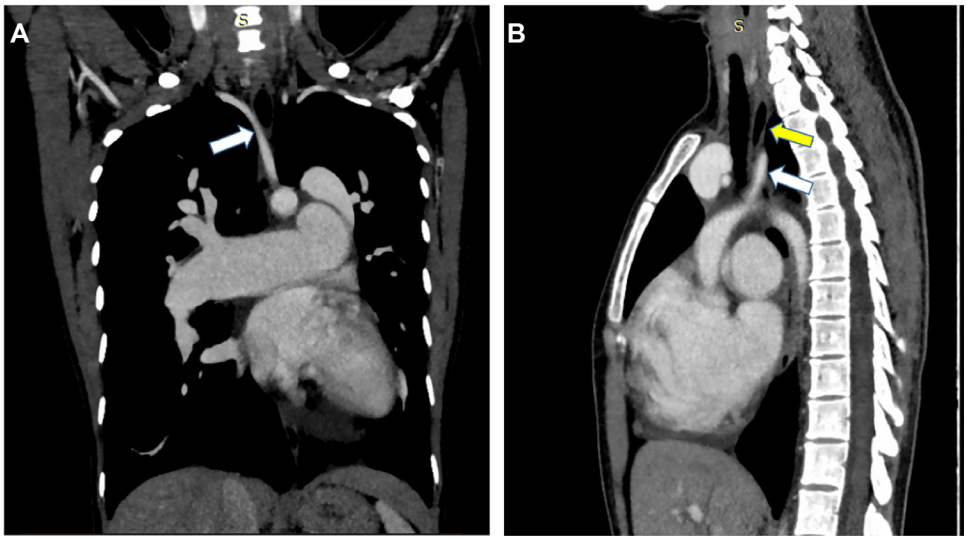

Fig. 8 – (A and B) Anomalous course of the aberrant right subclavian artery posterior to the esophagus (White Arrow: aberrant right subclavian artery (ARSA), Yellow Arrow: Esophagus).

resolution and field of view. In such cases, conventional angiography remains the gold standard despite its risks, with computed tomography emerging as a valuable tool for detecting subtle venous anomalies [7].

We also discovered an incidental finding of an aberrant right subclavian artery (ARSA) with a retroesophageal course. ARSA is a relatively rare [8] anomaly, occurring in 0.16%-4.4% of the general population, with a higher incidence in females. In patients with congenital heart defects, the incidence is around 3%, while it is much higher in those with Down syndrome [9]. There are no known studies linking PAPVC with ARSA. The retroesophageal ARSA can be vulnerable to compression and pressure necrosis from nasogastric or endotracheal tubes, potentially leading to arterio-esophageal fistula formation. However, our patient experienced no complications.

Conclusions

Partial anomalous pulmonary venous connections (PAPVC) are uncommon congenital anomalies that may be detected incidentally or present with variable symptoms, depending on the severity and associated defects. Advanced imaging techniques like MDCTA and MR angiograms provide excellent 3D visualization, aiding in diagnosis and management planning. The morphological variability and associated congenital anomalies significantly influence morbidity and mortality, making early and accurate diagnosis essential for effective treatment and decision-making.

Patient consent

Written informed consent was obtained from the patient for the case report "Partial anomalous pulmonary venous connection and an incidental aberrant right subclavian artery: A rare case report".

REFERENCES

[1] A Snellen B.H., Van Ingen H.C., M Hoefsmit E.C. *Patterns of anomalous pulmonary venous drainage*. http://circ.ahajournals.org/.

[2] Kasahara H, Aeba R, Tanami Y, Yozu R. Multislice computed tomography is useful for evaluating partial anomalous pulmonary venous connection. J Cardiothorac Surg 2010;5(1). doi:10.1186/1749-8090-5-40.

[3] Stiver C, Ball MK, Cua CL. Pulmonary Venous Anomalies. InPediatric Cardiology: Fetal, Pediatric, and Adult Congenital Heart Diseases, 6. Cham: Springer International Publishing; 2024. p. 1377–419.

[4] Wakamatsu H, Satokawa H, Nagata E, et al. An integrated treatment for isolated partial anomalous pulmonary venous connection with severe pulmonary hypertension. J Pediatr Cardiol Cardiac Surg 2024;8(2):70–6.

[5] Jin C, Wu Y, Wang Z, Liu X, Wang Q. Isolated partial anomalous pulmonary veins: a 10-year experience at a single center. J Surg Res 2024;298:63–70.

[6] Hegde M, Manjunath SC, Usha MK. Isolated partial anomalous pulmonary venous connection: ddevelopment of volume overload and elevated estimated pulmonary pressure in adults. J Clin Imaging Sci 2019;9(29):1–6. doi:10.25259/JCIS-8-2019.

[7] Deshpande AA. Imaging in Total Anomalous Pulmonary Venous Connections and Partial Anomalous Pulmonary Venous Connections. In: Case-based Atlas of Cardiac Imaging. Singapore: Springer Nature Singapore; 2024. p. 133–46.

[8] Tyczyński P, Michałowska I, Wolny R, et al. Left aberrant subclavian artery. Systematic study in adult patients. Int J Cardiol 2017;240:183–6. doi:10.1016/j.ijcard.2017.04.052.

[9] Natsis K, Didagelos M, Gkiouliava A, Lazaridis N, Vyzas V, Piagkou M. The aberrant right subclavian artery: cadaveric study and literature review. Surg Radiolog Anatom 2017;39(5):559–65. doi:10.1007/s00276-016-1796-5.